legs was beneficial, and he was able to walk occasionally without assistance.

Autonomic function tests were performed after weaning from mechanical ventilation.1 A catheter was inserted into the left radial artery, and vital signs were monitored. Head up tilting to a 60° head and a blood pressure of fall from 134/99 to 51/48 mm Hg. The basal plasma noradrenaline concentration was normal. A rise of plasma vasopressin in response to hypotension was absent, suggest- ing involvement of the efferent baroreflex pathway. There was no overshoot with Valsalva’s manoeuvre, which suggested insufficient baroreflex function. The absence of a blood pressure rise to the cold pressor test suggested involvement of the efferent sympathetic system. A noradrenaline infusion test disclosed sympathetic supersensi- tivity. No ß-1 supersensitivity in response to an isoprenaline infusion test was seen.2 Increase of his heart rate from 90 to 109 per minute in response to an atropine test showed that the vagal efferents were not completely involved. The results of the remaining autonomic system.

The control of blood pressure after pos- tural change is mediated by the baroreflex system. Failure of this reflex function can be due to a lesion anywhere in this reflex path- way. The baroreceptors, originating from the carotid sinus and aortic arch, travel via the glossopharyngeal and vagal nerves. Most of these baroreceptor afferents terminate in the nucleus tractus solitarius, the major nerve relay of the baroreflex.3,4 In addition, the nucleus truc- tus solitarii not only regulates baroreceptor pathways but also is involved in the central regulation of respiration and swallowing.5 The glossopharyngeal and vagal nerves also convey the sensory impulses from the pharynx, larynx, and the respiratory system to the nucleus tractus solitarii, which contains specific centres for respiration and swallow- ing.6 Our patient with severe orthostatic hypotension, dysphagia, lingual atrophy, and temporary failure of automatic respiration, but had no pyramidal signs, sen- sory ataxia, cerebellar ataxia, or a uro- genic bladder. Although we cannot discuss the relation of a non-metastatic autonomic neuropathy, neither spinal nor peripheral nerve lesions, as we found. Investigation of the somatic and autonomic functions sug- gested a medullary lesion. Severe orthostatic hypotension, the absence of overshoot of Valsalva’s manoeuvre, a negative response to the cold water test, and a positive response to the atropine test are consistent with an incomplete failure of the baroreflex arc. Although it is difficult to determine the defi- nite lesion responsible for the failure of the baroreflex arc, a tumour located in the dor- sal medulla may cause impairment of the nucleus tractus solitarii, the dorsal vagal nucleus, and the hypoglossal nucleus, which are involved in close proximity in the dorsal medulla.7

In conclusion, a localised lesion of the dorsal medulla as reported in this case may result in severe orthostatic hypotension, bul- bar paralysis, and automatic respiration in the absence of pyramidal or sensorimotor signs in the limbs.

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Tardive dystonia after neuroleptic treatment of Tourette’s syndrome

We report a patient with Tourette’s syn- drome who developed tardive dystonia mani- fest by a task specific, action induced oromandibular dystonia after 15 years of treatment with haloperidol. A 28 year old man with Tourette’s syn- drome since the age of seven had been treated with haloperidol (2–4 mg daily) with good effect. At age 19, vocal tics included eye blinking, facial grimacing, shoulder shrugging, head and neck movements, humming and squeaking noises, and occasional snapping-shut jaw movements. Obsessive-compulsive features included repetitive touching and counting behaviours. At the age of 28, he developed severe dystonic movements of the jaw, mouth, and tongue for the first time, which were activated by speaking. Immediately on beginning to speak he displayed forceful dys- tonic jaw opening and buccolingual move- ments associated with a pronounced dysarthria. He had no history of tic disorder. Examination was normal. He was able to control his dystonia by speaking with a finger or pencil clenched between his teeth. The mouth movements were absent at rest and were not activated by eat- ing or non-verbal jaw movements. There were also mild choreic movements of the hands and fingers. After stopping haloperi- dol there was transient exacerbation of jaw movements which lasted for several weeks followed by persistent speech activated jaw opening oromandibular dystonia, which remained unchanged for the next 10 months. Eye blinking and cervical tics exac- erated when haloperidol treatment was stopped, and improved with clonazepam with partial benefit. Benztropine and tetra- benzine were unhelpful for the oro- mandibular dystonia but beginning 10 months after onset there was slow improve- ment followed by complete resolution of the jaw dystonia, which has now been absent for 18 months.

Tardive dystonia and tardive dystonia have only rarely been reported in patients with Tourette’s syndrome after chronic neurolep- tic treatment.1 Our patient developed an action induced oromandibular dystonia after 15 years of treatment with haloperidol. Diagnosis of tardive dystonia occurring on a background of Tourette’s syndrome may be difficult. In this case, the patient’s dystonia differed from dystonic tics sometimes associ- ated with Tourette’s syndrome in being highly localised, action induced, repetitive, patterned, not suppressible, and unaccom- panied by a urge to move. 1 His severe dystarthria was also more consistent with oromandibular dystonia than a dystonic jaw tic. Because cranial and cervical dystonia have been reported in a few patients with tic disorders not exposed to neuroleptic drugs,1 the possibility of a coincident pri- mary dystonia unrelated to neuroleptic drugs also deserves consideration. The his- tory of prolonged neuroleptic exposure, exacerbation of dystonia after stopping haloperidol, and disappearance of dystonia after 12 months of stopping haloperidol are clinical features much more indicative of tardive dystonia induced by neuroleptic drugs than spontaneous dystonia.

Although tardive dyskinesia has been uncommon in patients with Tourette’s syn- drome, vigilance for the appearance of new involuntary movements, and their resolution when they appear dystonic or dystonic rather than tic-like, is appropriate in the manage- ment of Tourette’s syndrome with neurolep- tic drugs.

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Should patients with central core dis- ease be screened for malignant hyperthermia?

Malignant hyperthermia is a potentially fatal condition of hypermetabolism in skeletal muscle triggered by certain anaesthetic agents. Although neuromuscular disorders may predispose to anaesthetic complications in their own right, only central core disease and malignant hyperthermia myopathy are associated with the change in muscle metabolic responses diagnostic for susceptibil- ity to malignant hyperthermia.1 Central core disease is characterised by weakness, wast- ing, and orthopaedic complications. Muscle fibres show round and angular oxidative- staining, known as cores. Malignant hyperthermia myopathy is a non-specific his- tological myopathy, sometimes with cores, occurring in certain anaesthetic reactive patients, susceptible to malignant hyperther- mia. Most susceptible patients show normal muscle histology. A few show central core disease or malignant hyperthermia myop- athy. Conversely, many patients with central core disease are susceptible to malignant hyperthermia. Indeed an assumption has grown that all patients with central core dis- ease are susceptible to malignant hyperther-
Clinical, in vitro contracture test, and histopathological data

<table>
<thead>
<tr>
<th>Case</th>
<th>Myopathic symptoms</th>
<th>Malignant hyperthermia</th>
<th>In vitro contracture test results</th>
<th>Percentage of affected fibres with central cores</th>
<th>Percentage of type 1 fibres</th>
</tr>
</thead>
<tbody>
<tr>
<td>I.1</td>
<td>Severe</td>
<td>Susceptible</td>
<td>119</td>
<td>58</td>
<td>98</td>
</tr>
<tr>
<td>I.2</td>
<td>Mild</td>
<td>Susceptible</td>
<td>120</td>
<td>54</td>
<td>95</td>
</tr>
<tr>
<td>I.3</td>
<td>Absent</td>
<td>Susceptible</td>
<td>121</td>
<td>49</td>
<td>96</td>
</tr>
<tr>
<td>II.1</td>
<td>Absent</td>
<td>Susceptible</td>
<td>122</td>
<td>49</td>
<td>96</td>
</tr>
<tr>
<td>II.2</td>
<td>Absent</td>
<td>Susceptible</td>
<td>123</td>
<td>0</td>
<td>0</td>
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<tr>
<td>III.1</td>
<td>Absent</td>
<td>Susceptible</td>
<td>124</td>
<td>25</td>
<td>100</td>
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<tr>
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<td>Susceptible</td>
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<td>126</td>
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<tr>
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<td>127</td>
<td>127</td>
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<tr>
<td>V</td>
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<td>Absent</td>
<td>Susceptible</td>
<td>129</td>
<td>129</td>
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</tr>
</tbody>
</table>

Clinical data indicate that whereas central core disease and malignant hyperthermia may be related conditions, they are not the same. A similar genetic locus, chromosome 19q13, has been linked to malignant hyperthermia, but the protein product has more than one function. Perhaps the location of the mutation and the genetic background of the individual dictate whether one or both conditions are lost. Our suggestion is that patients with central core disease may be susceptible to malignant hyperthermia by being exposed to agents that would normally not be fatal when reacting normally to one agent. That such conditions may be inherited as a genetic disease is suggested by the clinical data. The mother's case (II, 1) showed a fulminant disease. The sister of the proband (case 1), a 37 year old woman, died in family III was congenitally affected. Her mother (case 2), daughter (case 3), and son (case 4) were also affected. In family I, the proband had central core disease and was susceptible to malignant hyperthermia. In family V, the patient had central core disease histologically. She had esophageal dysmotility, ankylosing spondylitis, and weakness of the legs, with a classic histological picture of central core disease, but was negative for malignant hyperthermia. The study indicates that patients with central core disease should be screened for malignant hyperthermia. They should not be assumed to be susceptible. A significant number are negative for malignant hyperthermia and if they are assumed to be susceptible this will unnecessarily deny them potentially valuable anaesthesia. It is important to know the reliability of the in vitro contracture test results in this study. A collaborative study from all European centres has yet to be published. In this study, no clinically fulminant patient with malignant hyperthermia has tested negative and a review of 402 probands shows a good correlation between the test result and the severity of the clinical reaction. This is supported by the finding of 100% sensitivity and 78% specificity using the essentially similar North American protocol.
the others, was normal percentages of type I fibres, rarely any high levels (90%) or more seen in the other families.

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MATTERS ARISING

Multiple sclerosis in the north Cambridgeshire districts of East Anglia

The north Cambridge survey is a welcome addition to the United Kingdom series of prevalence studies. We agree that a multi-centre prevalence study would add to the epidemiological knowledge of multiple sclerosis. However, we cannot agree with the inclusion of “suspect cases” in their prevalence figures. As we have pointed out in a previous paper, the measurement of multiple sclerosis can be distorted by using ill defined criteria for measuring the disease. We contend that the Poser criteria alone (which do not contain a suspect category) should be used in measuring the prevalence of multiple sclerosis. For this reason, we used only the Poser criteria for the survey of west Sussex, and deliberately did not include a suspect category. As Poser himself says, “for the purposes of prevalence studies only the categories of clinically definite and clinically probable should be used; possible multiple sclerosis should never be included.”

Our concern is that a “suspect” category, which seemed to have been defined differently in the south and west of England (Cambridge and Southamptoon surveys), can lead to confusion in interpreting and comparing prevalence figures. This is because there are no clear criteria of what constitute so called “suspect” cases, and workers are free to use their own criteria. As Robertson says, the inclusion of a suspect category “introduces noise, and generally obscures the overall picture.” We agree with this, and argue that any cases that do not fall into the Poser criteria should be left as prevalence figures. To do so would introduce some clarity into what we are striving to measure. In view of future prevalence surveys should use the Poser criteria and not include “suspect” cases.

The Cambridge team suggest that the very presence of a latitudinal gradient within the United Kingdom has only recently been questioned. It is, in fact, a decade ago that Williams and McKeegan made the comment “we find no convincing evidence of a latitudinal effect in the United Kingdom.” A mortality study of multiple sclerosis in the United Kingdom found no gradient south of the North Sea border and discussed the possibility that the high, but diminishing, Scottish rates were artefactual. The most serious challenge to the latitudinal hypothesis appears in a letter in the BMJ in 1993; convincing argument was presented to show that the hypothesis was inconsistent with United Kingdom data.

So the challenge to Limburg’s hypothesis is not recent. What is recent is that most researchers in the field are coming to realise the weakness of the data on which the hypothesis was based.

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NOTICE

Announcement from the British Neuro-psychiatry Association: 1996 summer meeting

The 1996 Summer meeting will be held on 14-16 July at Robinson College, Cambridge. It will include topics on neuro-development, language, and the presentation of short scientific papers and single case videos by members. The Association’s AGM will be held on 16 July.

For further details of these meetings please contact: Sue Garratt, Administrative Assistant, BNPA, 17 Clocktower Mews, London N1 7BB. Telephone/Fax: 0171 226 5949.

For details of membership of the BNPA, which is open to medical practitioners in psychiatry, neurology, and related clinical neurosciences, please contact: Dr Jonathan Bird, Secretary, BNPA, Borden Neuropsychological Hospital, Stoke Lane, Stapleton, Bristol, BS16 1QT. Telephone: 01179 701212 ext 2925/2929 or Sue Garratt at the address given above.

CORRECTION


In the table, p223, CT localisation of patient B is Left frontal. NA [no abnormalities] The first sentence, left hand column p223 should read—Both sides. NA and CT showed abnormalities that were not in accordance with EEG findings.

BOOK REVIEWS

All titles reviewed here are available from the BMJ Bookshop, PO Box 295, London WC1H 9TE. Prices include postage in the United Kingdom and for members of the British Forces Overseas, but overseas customers should add £2 per item for postage and packing. Payment can be made by cheque in sterling drawn on a United Kingdom bank, or by credit card (Mastercard, Visa or American Express) stating card number, expiry date, and your full name.


The third edition of Sophie Levitt’s excellent book will be of interest and indeed is essential reading for anyone involved in the management of the cerebral palsies, including parents. A major theme throughout the book is the importance of collaboration with parents and the detailed section on practical procedures is written with parents as well as therapists in mind. Forwards to the second